A Case of Mycosis Fungoides and Lymphomatoid Papulosis Occurring Simultaneously in a Child

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ABSTRACT

Several studies have reported an association between lymphomatoid papulosis and other lymphomas, such as mycosis fungoides, Hodgkin's disease, and anaplastic large cell lymphoma. The association between lymphomatoid papulosis and mycosis fungoides has been reported to be between seven and 39 percent. Although a relationship is acknowledged between lymphomatoid papulosis and mycosis fungoides, our understanding is limited. The authors report a case of mycosis fungoides and lymphomatoid papulosis in a child.
(*J Clin Aesthet Dermatol.* 2012;5(11):46–48.)

An 11-year-old girl presented with a six-year history of asymptomatic, light patches on her face, arms, and legs. The patches initially faded in the winter months, but more recently have remained fixed. The patient also reported a three-year history of pruritic papules on her left forearm and wrist, a rash that started during a vacation in Jamaica. Topical corticosteroids and tacrolimus 0.1% ointment have reduced the erythema and scale on her face, but have not helped the pruritic papules on her left forearm. Review of systems was otherwise negative. The patient's past medical history has otherwise been unremarkable.

On physical examination, there were several pink to hyperpigmented papules on her left dorsal wrist (Figure 1). There were also multiple large, hypopigmented patches on her arms and legs (Figure 2). In addition, there were hypopigmented and erythematous patches with slight scale on the bilateral cheeks.

Four skin biopsies were taken. The left forearm sample showed a wedge-shaped, dense perivascular and interstitial infiltrate of lymphocytes with CD30(+) cells and a few CD20(+) cells. T-cell gene rearrangement studies were positive for distinct monoclonal peaks (Figure 3). The right arm biopsy displayed mild psoriasiform hyperplasia with lymphocytic epidermotropism, mild spongiosis, and necrotic keratinocytes. Few CD30(+) cells and CD20(+) cells were present. T-cell gene rearrangement studies were positive and identical to the left forearm sample (Figure 4). The left anterior thigh sample showed a superficial perivascular lymphoid infiltrate with mild epidermotropism. There were few CD30(+) cells with no CD20 positivity. T-cell gene rearrangement studies were predominantly negative, yet suspicious amplicons with identical size to the first two specimens were present. The left dorsal wrist sample displayed a perivascular infiltrate of lymphocytes accompanied by interstitial neutrophils and mild spongiosis. Some of the lymphocytes were large and pleomorphic with a moderate number of CD30(+) cells and few CD20(+) cells. T-cell gene rearrangement studies were suspicious for a positive and partial match to the left forearm and right arm samples. In addition, flow cytometry was performed and was within normal limits and an antinuclear antibody (ANA) was negative.

Thus, the authors diagnosed the patient with mycosis fungoides (MF)—patch stage—and lymphomatoid papulosis (LyP)—type A. She was treated with narrowband ultraviolet B (UVB) phototherapy (28 treatments) that has led to repigmentation of all of the hypopigmented areas, but has not improved the pruritic lesions on her left wrist. The plan is for her to continue narrowband UVB phototherapy for another 12 treatments.

DISCLOSURE: The authors report no relevant conflicts of interest.
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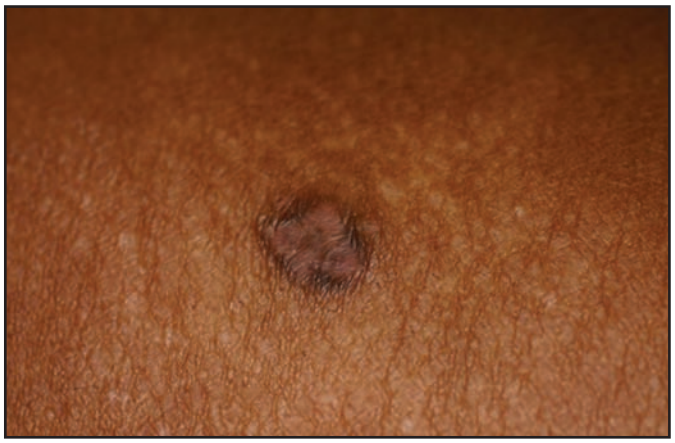

Figure 1. Pink, hyperpigmented papule on the left dorsal wrist

Figure 2. Multiple large, hypopigmented patches on the patient's arms

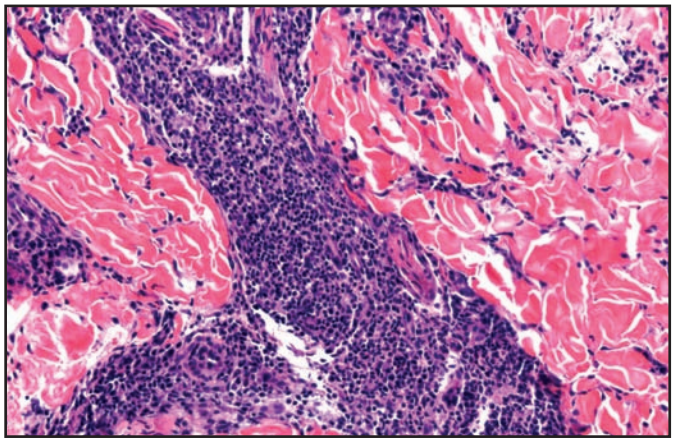

Figure 3. A high power view of the biopsy taken from the patient's left forearm demonstrating a wedge-shaped, dense perivascular and interstitial infiltrate of lymphocytes with many CD30(+) cells

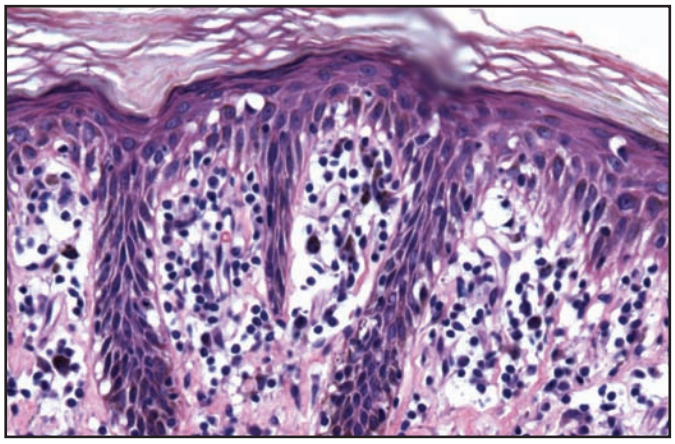

Figure 4. A low power view of the biopsy taken from the patient's right arm illustrating mild psoriasiform hyperplasia with lymphocytic epidermotropism, mild spongiosis, and necrotic keratinocytes.

DISCUSSION

Mycosis fungoides is a CD4(+) T-cell lymphoproliferative disorder that has an indolent course in its early stages. Histologically, a superficial lymphocytic infiltrate is seen abutting the dermoepidermal junction. Lymphocytes have hyperchromatic, cerebriform nuclei. Minimal spongiosis is associated with epidermotropism and Pautrier microabscess formation. Hypopigmented MF is seen more often in the younger population and is the most common form of cutaneous T cell lymphoma in children.[6,7] The incidence of MF in patients younger than 20 years of age is about 0.5 to five percent of the total incidence of MF.[8] Treatment options for children with hypopigmented MF are generally the same as that of classic MF. For patch-stage disease, topical corticosteroids, retinoids, or phototherapy are first-line treatments.

In the past, there was an ongoing debate over the prognosis of MF in children versus adults. Fink-Puches et al[9] revealed that 17 of 24 MF patients under the age of 20 years did not experience progression of the disease or death.[9] Wain et al[10] also reported that the prognosis of MF in children under the age of 16 did not show significant differences when compared to adult-onset MF.[10] Conversely, Peters et al[11] in 1990 found in his study that three out of five MF children had metastasis to the lymph nodes and the prognosis was worse than that of adult-onset MF.[11] However, in recent years, studies have shown childhood MF has a good prognosis, but requires long-term treatment and follow up.

Lymphomatoid papulosis is a benign CD30(+) lymphoproliferative disorder that typically presents as recurrent crops of red-brown papulonodules with necrotic centers on the trunk or extremities.[12] Lesions usually self-heal in 4 to 6 weeks, leaving hyperpigmentation or atrophic scars. Histologically, a wedge-shaped inflammatory infiltrate with many CD30(+) lymphocytes is seen. Type A LyP has a background of histiocytes, eosinophils, and neutrophils. In type B LyP, epidermotropism is seen, resembling mycosis fungoides. Type C resembles type A, but with sheets of anaplastic large cells.[13] A recently proposed type D has marked epidermotropism of CD8(+) T cells.[13] Although benign, clonal T-cell gene rearrangement can be demonstrated in more than half of LyP cases.

LyP patients have a 10- to 20-percent increased risk of developing lymphoma, most commonly MF, primary cutaneous anaplastic large-cell lymphoma, and Hodgkin's disease. This is well documented in adults; however, this transformation rarely occurs in the pediatric population. There have only been two reported cases of lymphoma developing after a LyP diagnosis in a pediatric patient.[14,15] While LyP is a benign disease that does not require treatment, it can be associated with lymphoma, requiring close follow up. LyP treatment options include topical steroids, topical bexarotene, phototherapy, nitrogen mustard, methotrexate, and intralesional interferon.

Multiple studies have further shown how MF and LyP may be related by demonstrating identical clonal T-cell peaks on gene rearrangement polymerase chain reaction (PCR) studies. Zackheim et al[12] found that 21 (39%) of their LyP patients had co-existing MF lesions, and an identical clone was found in all seven patients in whom analysis was performed. A year later, Gallardo et al[16] published a study of 12 patients with co-existing LyP and MF, where T-cell clonality was identified in seven LyP lesions (58%) and six MF lesions (50%).[16] In every case demonstrating T-cell clonality, the MF and LyP lesions exhibited identical peak patterns via high-resolution PCR.[12,16]

In conclusion, the authors described a case of two rare lymphoproliferative disorders occurring simultaneously in a child. Overall, prolonged follow up of patients with MF and LyP reveals excellent prognosis with similar treatment modalities and a low mortality rate. Cutaneous recurrences are common, and although systemic spread is rare, it is possible. Awareness of the association between MF and LyP by clinicians and pathologists may lead to improved diagnosis, treatment, and follow up.

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